

Staged septation of double inlet left ventricle

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SUMMARY Complete septation of a double inlet left ventricle with left anterior subaortic outlet chamber was carried out successfully as a staged procedure during the first two years of life in a patient with severe pulmonary hypertension and an abnormal left atrioventricular valve. In contrast with isolated pulmonary artery banding, which rarely has led to a satisfactory septation or modified Fontan operation, this technique achieved good initial palliation and uncomplicated intracardiac repair. It should be considered for infants with univentricular atrioventricular connection, two atrioventricular valves, and excessive pulmonary blood flow.

Although theoretically attractive,¹ septation of the univentricular heart during infancy has been hindered by technical difficulties in securing the prosthetic patch and frequent postoperative heart block. Ebert recently demonstrated in five infants with "common ventricle" that two Teflon patches without a pressure difference across them could be positioned accurately with a few sutures, and that over a period of 5-8 months they became firmly adherent to the endocardium.² This avoided injury to the conduction system and created a stable base to which a third patch was attached, completing the prosthetic septum. The following case illustrates successful extension of this concept to a patient with double inlet left ventricle and subaortic outlet chamber.

Case report

A baby girl weighing 5.5 kg was first seen at six months of age during emergency hospital admission for a chest infection. She was said to have been normal at full term delivery in Iran but had failed to thrive. A heart condition was diagnosed when she was three months old.

On examination, the infant was pyrexial, tachypnoeic, and mildly cyanosed with clubbing. Bounding pulses at a rate of 160 beats per minute were present throughout. Enlargement of the heart with a thrill along the left sternal edge was detected by palpation. In addition to a loud systolic murmur, auscultation disclosed a single, accentuated, second heart

sound. The liver was enlarged, and both lungs had diffuse crepitations. Chest x-ray showed cardiomegaly with plethoric lung fields. The electrocardiogram had a vertical axis and dominant left ventricular forces. Cross sectional echocardiography showed a large chamber of left ventricular morphology that received both atrioventricular valves and gave off the pulmonary artery. The aorta arose from a left-sided outlet chamber. The left atrium was dilated and the left atrioventricular valve appeared to be thickened.

The patient's heart failure responded to treatment with digoxin and diuretics, and gram negative pathogens in the urine and sputum were cleared with antibiotics, such that cardiac catheterisation could be performed ten days after the start of treatment. This confirmed the echocardiographic diagnosis and showed pulmonary hypertension, with pressures in the ventricle and ascending aorta being 83/6 mm Hg and 83/49 mm Hg, respectively, and in the main pulmonary artery 64/31 mm Hg. Mean left atrial pressure was 15 mm Hg compared with 5 mm Hg in the right atrium, and pulmonary vascular resistance was 3.2 units.m². Pulmonary flow was two and a half times systemic, and systemic arterial saturation was 91%.

Although cardiac failure was controlled, the infant failed to gain weight and remained ill. She was admitted to hospital with repeated chest infections. A second catheterisation at 14 months showed virtually equal systemic and pulmonary blood flow, with a pulmonary resistance of 10.9 units.m² in air, falling to 2.1 units.m² in 100% oxygen.

First stage septation and pulmonary artery banding were performed when she was 15 months old and

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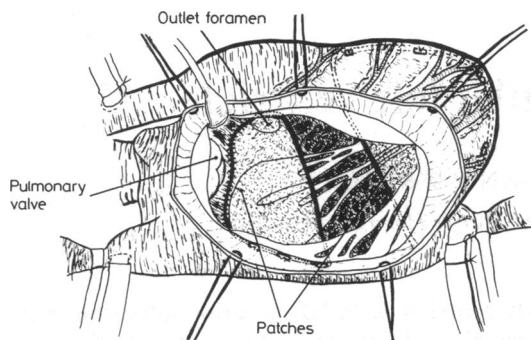


Fig 1 Surgical view of the first stage operation. The ventricle is exposed through the right atrium by retraction of the right atrioventricular valve. The left atrioventricular valve and outlet foramen lie behind the upper Teflon patch, while the lower patch separates the tensor apparatus of left and right atrioventricular valves.

weighed 6.7 kg. The left atrioventricular valve, inspected on cardiopulmonary bypass through an enlarged patent foramen ovale, was found to be thickened but adequate in size. Exposure was set up through the right atrioventricular valve, and a patch of Teflon felt was positioned between the two inlet valves, the outlet foramen, and the pulmonary artery with a continuous suture (fig 1). In the region of the conduction system, stitches were taken inside the upper margin of the outlet foramen.³ A second, triangular patch was attached at the apex of the ventri-

cle with interrupted mattress sutures supported by Teflon pledgets outside the heart. After the patient had come off bypass the diameter of the pulmonary artery was reduced to about 7 mm by banding. Together, the patches constituted approximately 75% of a ventricular "septum".

Cardiac function was excellent in the immediate postoperative period while the patient was on 0.1 µg/kg/min isoprenaline. Systolic arterial blood pressure was 100–110 mm Hg and she was in sinus rhythm. The patient later suffered transient supra-ventricular tachycardia which was controlled by rapid atrial pacing, and fever with seizures, attributed to pseudomonas infection of the sputum and urine. Despite this she was extubated on the third postoperative day and made an otherwise uneventful recovery.

During the next six months she grew well without signs of heart failure or respiratory infection. Cardiac catheterisation five months after operation showed satisfactory banding of the pulmonary artery, with a distal pressure of 22/11 mm Hg and pulmonary resistance of 2.9 units.m². The patches were both in a good position, but there was an 8 mm Hg gradient across the outlet foramen, which appeared to be restrictive on both echocardiography and angiography (fig 2).

At the time of her second open heart operation, in February 1986 the patient was 22 months old and weighed 9.6 kg. The previously inserted Teflon was found to be completely endothelialised and firmly

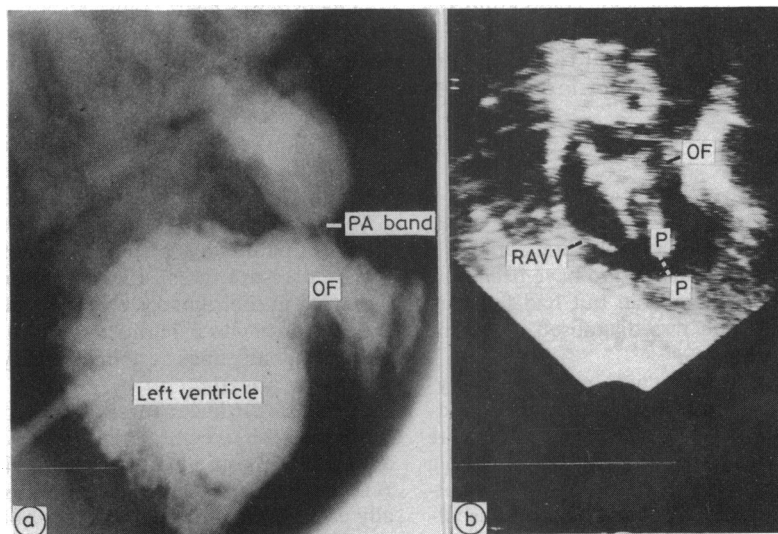


Fig 2 Left ventricular angiogram (a) and echocardiogram (b) after first stage septation and pulmonary artery banding. The dotted line between P and P indicates position of third patch, which later completed the ventricular partition. OF, outlet foramen; PA, pulmonary artery; RAVV, right atrioventricular valve; P, septation patches.

adherent to the ventricular endocardium. The outlet foramen was enlarged by excision of a wedge of muscle from the posterior inferior trabecular septum, the pulmonary artery was debanded, and septation was completed by transatrial insertion of a third Teflon patch.

Postoperatively the patient remained in sinus rhythm. A transient increase of left atrial pressure from 12 mmHg to 20 mmHg was treated by inotropic support (5 µg/kg/min dobutamine) and afterload reduction (glyceryl trinitrate). Extubation was performed uneventfully on the second day after operation, and the patient was discharged from hospital one week later. Doppler echocardiography before discharge showed no clinically important gradient across the pulmonary artery or outlet foramen or any shunt across the prosthetic septum. There was a trace of regurgitation through the left atrioventricular valve. Ventricular contractility was good and movement of the septal patches was minimal.

Discussion

Prognosis is generally poor in patients with double inlet ventricle. Of those presenting in the first year of life, successful "correction" by ventricular septation or a modified Fontan operation was achieved in <2% of one series⁴; while in another, half of those who survived to childhood and did not undergo surgery died within fourteen years of diagnosis.⁵ Appropriate surgical intervention has been shown to improve both the length and quality of life for these patients,¹⁻⁶ but management of those with increased pulmonary blood flow continues to be difficult.

Many in this subgroup develop cardiac failure and pulmonary hypertension with respiratory infections and failure to thrive early in life. Banding of the pulmonary artery can be done with a low operative risk⁷ and ten year actuarial survival is 74%⁶; but even after haemodynamically effective banding, pulmonary vascular structure may remain abnormal and later prejudice the outcome of a Fontan procedure.^{8,9} Although subtle structural anomalies and even moderately increased pulmonary arterial resistance are tolerable with the septation operation, loss of ventricular compliance, outlet foramen obstruction, small ventricular size, and intrapericardial adhesions all significantly increase early mortality after septation¹⁻⁶ and may be caused or accentuated by banding.^{10,11}

Insertion of two patches within the ventricle combined with pulmonary artery banding for a limited period appears to be a safe and effective means of palliation that also facilitates definitive septation. Our patient differed from those reported by Ebert in having a more common type of double inlet ventricle

(that is both atria were connected to a dominant left ventricle with a left sided, anterior rudimentary right ventricle), raised pulmonary vascular resistance, and an abnormal left atrioventricular valve. These latter two factors almost certainly would have precluded a successful Fontan operation in the future. None the less, both stages of septation were followed by a relatively uneventful postoperative convalescence. In retrospect, it would have been better to perform the first stage septation and pulmonary artery banding after the initial investigation, because the operative risk increased with the subsequent considerable rise in pulmonary vascular resistance. Because the hazards of cardiopulmonary bypass may be greater in very young infants,¹² it would be reasonable to carry out isolated pulmonary artery banding if operation were required during the first two or three months of life, particularly for associated coarctation of the aorta. The initial stage of septation could then be undertaken between three and six months of age. To delay protection of the pulmonary vasculature for more than eight months probably increases the likelihood of permanent changes in the lungs,¹³ and accordingly palliation should start before this time.

There are several important unanswered questions about this procedure. It is not known whether outlet foramen obstruction will recur. The fate of atrioventricular valves in a growing heart whose inlet to apex length is fixed, the incidence of thromboembolic complications during patch endo-thelialisation, and the late results as regards ventricular function and cardiac rhythm are uncertain. Early survival and functional results have been encouraging, however, and they support the continued development of techniques for septation in infants with suitable ventricular morphology and excessive pulmonary blood flow.

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